Shunt age-related complications in adult patients with suspected shunt dysfunction. A recommended diagnostic workup

Spirig, José M ; Frank, Melanie N ; Regli, Luca ; Stieglitz, Lennart Henning

Abstract: BACKGROUND: Patients admitted for suspicion of shunt dysfunction (SD) often show unspecific symptoms and require time-consuming, expensive and even invasive diagnostics involving significant radiation exposure. The purpose of this retrospective study was to analyse the current diagnostic procedures and to propose a process optimisation. METHOD: As all patients admitted for suspicion of SD receive imaging studies, we searched for adult patients receiving neuroimaging in the period from January 2010 to July 2013, analysing referring diagnosis, clinical signs, products, diagnostic process and final diagnosis. Recursive partitioning was used to define time intervals for differentiating types of SD. RESULTS: A total of 148 patients, aged 18-89 (mean, 54) years, were studied. Forty-two percent were referred by a hospital or rehabilitation centre, 30% by general practitioners and 24% were self-referrals. The admission diagnosis was in the majority "shunt dysfunction" only. Further differentiations were rarely made. An SD was confirmed in 46% of the patients. In 17%, the symptoms were based on another cause and in 37% they could not be clearly attributed to any specific disorder. Abdominal dislocations (2%) and shunt infections (5%) were found within the first 6 months. Over- (3%) and under-drainage (14%) were the most frequent complications during the first 4 years. Disconnections (13%) occurred generally 4 years or more after implantation. Only shunt obstruction (9%) showed no temporal pattern. CONCLUSIONS: Symptoms of SD remain mostly unspecific. This study showed that the type of SD depends on the time interval from implantation. We propose a workup strategy for patients with SD based on the temporal profile.

DOI: https://doi.org/10.1007/s00701-017-3237-6

Shunt-age related complications in adult patients with suspected shunt dysfunction. A recommended diagnostic workup

Authors:

*José M. Spirig, MD,[1] *Melanie N. Frank, MB[2], Luca Regli, MD[1], Lennart H. Stieglitz, MD[1]

[1] Department of Neurosurgery, University of Zurich and University Hospital Zurich, 8091 Zurich, Switzerland

[2] Faculty of Medicine, University of Zürich Switzerland

* These authors contributed equally to this work

[a]Correspondence should be addressed to:

Lennart Henning Stieglitz MD

University Hospital Zürich, Neurosurgical Department, Zürich, Switzerland

Email: lennart.stieglitz@usz.ch

Phone number: +41 44 255 99 05

Fax number: +41 44 255 45 05

ABSTRACT

Background

Patients admitted for suspicion of shunt dysfunction (SD) often show unspecific symptoms and require time consuming, expensive, and even invasive diagnostics involving significant radiation exposure. The purpose of this retrospective study was to analyze the current diagnostic procedures and to propose a process optimization.

Methods

As all patients admitted for suspicion of SD receive imaging studies, we searched for adult patients receiving neuroimaging in the period from January 2010 to July 2013 analyzing referring diagnosis, clinical signs, products, diagnostic process and final diagnosis. Recursive partitioning was used to define time intervals for differentiating types of SD.

Results

148 patients aged 18 – 89 (mean: 54) years were studied. 42% were referred by a hospital or rehabilitation center, 30% by general practitioners and 24% were self-referrals. The admission diagnosis was in the majority "shunt dysfunction" only. Further differentiations were rarely made.

A SD was confirmed in 46% of the patients. In 17%, the symptoms were based on another cause and in 37% they could not be clearly attributed to any specific disorder.

Abdominal dislocations (2%) and shunt infections (5%) were found within the first 6 months. Over- (3%) and underdrainage (14%) were the most frequent complications during the first 4 years. Disconnections (13%) occurred generally 4 years or more after implantation. Only shunt obstruction (9%) showed no temporal pattern.

Conclusion

Symptoms of a SD remain mostly unspecific. This study showed that the type of SD depends on the time interval from implantation. We propose a work-up strategy for patients with SD based on the temporal profile.

Key Words: shunt dysfunction, shunt complication, shunt malfunction, shunt failure, ventriculoperitoneal shunt, cerebrospinal fluid shunt

INTRODUCTION

Complications in patients with CSF shunts are very common. About 18% to 32% of the shunts need to be revised within 5 years after the implantation, for pediatric patients even up to 50% within the first year[8, 10]. Major complications such as obstructions, disconnections, breaks, leakage or infections, have a yearly incidence of up to 60%. [4, 7]

Thus, patients with implanted CSF shunts are frequently admitted to a neurosurgical department for suspected shunt dysfunction (SD).

Unfortunately, SD is difficult to verify because the symptoms are unspecific and individual. Drowsiness, headache, vomiting and lethargy, that are predictive of acute shunt malfunction are non-specific [1, 5]. Furthermore, they are depending on the rapidity of onset of the condition. For all these reasons practitioners have a tendency to refer the patients frequently. Patients with shunts are probably admitted for evaluation of a SD more often than necessary.

Routine use of radiographic imaging in the evaluation for possible SD include mostly a CT scan of the head and several radiographs of the skull, the chest and the abdomen. Excessive use of this shunt series produces unnecessary costs and exposure to radiation.

Moreover, many referrals of patients with suspected shunt failure are poorly founded and therefore the suspicion is often refused. These patients are unnecessarily exposed to invasive investigations.

The aim of this study was to identify clinical features that would lead to a more focused and thus less invasive diagnostic workup. We retrospectively reviewed clinical presentation and diagnostic workup in patients referred to our hospital for suspected SD. We found that different shunt dysfunctions had certain accumulation periods after shunt surgery. This fact has not been described in the literature so far. According to these findings we suggest a new diagnostic paradigm.

MATERIAL AND METHODS

Ethical approval was obtained before data acquisition from the local ethics commission (KEK-ZH 2013-0587).

As all patients admitted for suspicion of SD regularly receive image-based diagnostics, we searched for patients with CSF shunts in the order mask of the departments of diagnostic radiology and neuroradiology in the period from January 2010 through July 2013.

The medical records of each subject found were reviewed and patients with suspicion of SD were selected. The following data were collected: referring physician, referring diagnosis, clinical signs, type of implanted shunt system, time since implantation, diagnostics performed at our institution and our final diagnosis. Eleven final diagnoses were differentiated. SD were distinguished into abdominal dislocation, under- and over-drainage, blocking, shunt infection, disconnection, kinking and laceration of the shunt. Under-drainage was differentiated from blocking by considering under-drainage a problem of the valve pressure adjustment whereas blocking had to be solved by a shunt valve replacement. Over-drainage was also defined as a valve pressure misadjustment. Diagnoses were confirmed by symptoms relieve after correction of the underlying problem.

The diagnosis of "others" was given when there was not a shunt related reason for the symptoms found and the patient recovered from an alternative diagnosis such as appendicitis, epilepsy or musculoskeletal disorder. "No diagnosis", which resulted in no neurosurgical intervention, was considered confirmed if the patient made a recovery in the further course and did not re-present with further symptoms and signs suggesting shunt malfunction or any other diagnosis.

Statistical Analysis

Data were analyzed and graphs were created using JMP statistics program (JMP 10.0, SAS Campus Drive, Cary, NC, USA).

Descriptive statistics were used to describe the study population, referral patterns, symptoms, diagnostics performed and final diagnosis. The Student t-test was used to compare means of metric variables whenever appropriate. Values are expressed as the mean and the 95% Confidence Interval (CI), unless indicated otherwise. $p < .05$ was considered statistically significant. Recursive partitioning analysis was used to create a decision tree. Results of the Receiver Operating Characteristic (ROC) Analysis are indicated as Area under the ROC curve (AUROC).

RESULTS

- **Demographics**

148 adult patients were included in the study, of whom 79 (53%) were women. The mean age of patients was 54 years with a range from 18 to 89 years. A bimodal age distribution was observed with a peak before 30 years and the other around 80 years of age.

The latter agrees to the rising prevalence of idiopathic normal pressure hydrocephalus, post-hemorrhagic hydrocephalus and hydrocephalus due to tumors in this age cohort. However, the former represents the expected even larger group of children with hydrocephalus becoming adult. Children were not included in the study, therefore the first peak only shows a small portion of what is to be expected.

- **Referral pattern**

The referring diagnosis was in almost all cases "shunt dysfunction". Further differentiations of the dysfunction were rarely made. Nearly three-quarters of patients were referred by either a general practitioner (GP) (n=45, 30%) or clinical institutions as hospitals and rehabilitation facilities including other specialists (n=65, 44%). One quarter (n=36, 24%) were patient self-admissions. (Table 1)

- **Shunt valves**

The two most frequently implanted valve types were Codman Hakim programmable valves ™(n=65, 44%) and Sophysa Mini SM8™ (n=60, 41%). It should be mentioned that the time of initial shunt implantation ranges from 1973 to 2013 (interquartile range: 2002-2010) and most of the patients in this study cohort were operated at our hospital. The two valves mentioned above, have been used most commonly in this time period.

- **Diagnoses**

In 68 (46%) of the cases the diagnosis of a SD could be confirmed and an appropriate therapeutic procedure could be performed. In 25 (17%) another cause of the symptoms could be found such as a musculoskeletal cause or a psychological factor (such as stress disorder). In 55 (37 %) cases the cause of the symptoms could not be identified, but a SD was ruled out. The most frequent reason for SD was under-drainage with a total of 20 patients (14%). Other recurrent causes included shunt disconnection, kinking or laceration (n=19, 13%) and blockage (n=13, 9%), shunt infection (n=8, 5%), over-drainage (n=5, 3%) and abdominal dislocation (n=3, 2%). (Figure 1)

- **Symptoms leading to admission**

Figure 2 summarizes the most common diagnoses and the respective symptoms that led to suspicion of a SD.

The leading symptom for over-drainage was headache which was present in all 5 patients.

Other common symptoms were nausea and vomiting, gait disorder, visual disorder or fatigue. Each occurred in two (40%) of these patients.

The most common symptom in under-drainage was gait disorder in 10 of 20 cases (50%). Eight patients (40%) had headaches, nine (45%) newly occurred neuropsychological deficits and six (30%) nausea and vomiting. Less frequent were incontinence and visual disorders in five (25%) and four cases (20%) respectively.

Neuropsychological deficits were defined as attention, memory or executive function impairment.

The most frequent symptom in shunt obstruction was headache in 8 of 13 patients (61.5%). Second most common were increasing gait disorder and nausea or emesis in four cases (31%) each.

Gait disorder and newly occurred neuropsychological deficits in 10 of 19 patients (52.6 %) as also urine incontinence (n=6, 32%) were most prominent in shunt disconnection.

In the 8 patients with shunt infection various symptoms such as headache (n=3, 37.5%), nausea/emesis (n=2, 25%) and reduced vigilance (n=3, 37.5%) could be observed.

All of the 3 patients with abdominal shunt dislocation had a palpable abdominal cyst and this symptom appeared only in abdominal shunt dislocation. It was therefore highly predictive for this diagnosis.

No other specific symptom or symptom combination could be found to differentiate between each diagnosis.

- **Diagnostic procedures performed**

Table 2 shows the frequencies of the invasive diagnostic procedures performed. A CT scan of the head was by far the most frequent procedure (n=128, 86%). 58% of all admissions had even an additional CT with contrast agent. More than half of the patients had an x-ray of either cranium, thorax, abdomen or all three. A shunt tap was only done when there was a strong suspicion of a shunt-infection (11%).

- **Relation of Shunt age and Type of complication**

We found no difference in shunt age nor in distribution of occurrence between the group with real SD, the group with another diagnosis than SD and the group without diagnosis. In contrast, we found differences between each type of SD. The overall mean time interval from date of implantation till occurrence of a complication was 5.4 years (95% CI: 4.4-6.5). Of all SD 36 (53%) presented before 3 years.

Abdominal dislocation was the earliest occurring complication with a mean time of 1.2 months (95% CI: -2.6-4.97). It was only reported in the first 6 months after a shunt implantation or revision. 50% of shunt infections occurred within the first 2 months but could also rarely be found years later (Mean: 2.3 years; 95% CI: -1.76-6.4). Over-drainage (Mean: 1.14 years; 95% CI: 0.003-2.277) and under-drainage (Mean: 2.43 years; 95% CI: 1.6-3.3) were common after one or two years. Late complications were disconnections, kinking or laceration (Mean: 9.86 years; 95% CI: 7.0-12.7) and blocking of the shunt (Mean: 8.0 years; 95% CI: 3.4-12.7). However, blocking of the shunt appeared in a constant frequency after 6 months (Figure 3). According to the time of occurrence it was possible to organize the complications into an early and late complication group. There was a statistically significant difference in time of occurrence between the early complication group (Mean: 2.03 years; 95% CI: 0.4-3.7) including abdominal dislocation, shunt-infection, over-drainage and under-drainage compared to the late complication group (Mean: 9.1 years; 95% CI: 7.4-10.9) including disconnections, kinking or laceration and blocking of the shunt ($p<0.0001$).

Based on these findings we created a decision tree using recursive partition analysis (Table 3 and Figure 4). Since the decision tree only contributes to discriminate between real shunt dysfunctions, diagnoses other than real shunt dysfunctions were excluded for this analysis.

The first partition was made at 6 months after implantation. During this time section abdominal dislocations (22%) and shuntinfections (44%) represent more than 60% of shunt dysfunctions. We therefore propose a clinical examination and an abdominal x-ray as first investigation. Less frequent are over- and under-drainage with 15% and 16% respectively. They are ruled out in a second step by a cranial CT. Eventually a shuntinfection is ruled out last by a shunt-tap since it is considered the most invasive procedure.

Between 6 months and 4 years over-drainage (13%), under-drainage (60%) and blocking of the shunt (22%) are the predominant entities. We therefore propose a cranial CT first.

After 4 years, disconnection, kinking or laceration are by far the most frequent SD (55%). This makes an x-ray of the complete shunt course as a first diagnostic procedure obligatory.

The performance of the partition analysis is summarized by the AUROC values for each type of SD using the mentioned time sections to discriminate between each type of SD (Table 4).

DISCUSSION

Admissions for suspected SD are very frequent. This study demonstrated, that 54% of all admissions don't have any shunt related problem but are exposed to radiating diagnostic procedures such as CT or X-rays in more than 85% and 55% respectively. In 17% of all admissions another health related problem such as a musculoskeletal or a psychological disorder is found and 37% remain unclear. Several reports also with similar high or even higher results are found in the literature. Cohen et al. [3] and Lehnert et al. [6] report, that as much as 80% and 77% of patients respectively won't have any neurosurgical intervention when they present for suspected SD. In contrast Barnes et al. [1] report that less than 30% are diagnosed as having normal shunt function after being thoroughly examined and Sribnick et al. [9] report up to 27% of patients admitted to the emergency department with suspected SD will not have any shunt revision.

The reason why so many normal functioning shunts are admitted is the fact that the symptoms remain unspecific and therefore it is difficult to clinically differentiate real SD from other disorders. On the other hand a real SD can be life threatening or have major neurological consequences including blindness.[2] This leads to a low-threshold referral behavior.

Several studies tried to identify specific symptoms or combination of symptoms which would reliably predict a SD. Barnes et al. [1] found drowsiness as the most predictive symptom for shunt block. Cohen et al. [3] found no association of any symptoms with radiological signs of shunt failure. Kim et al. [5] found lethargy and shunt site swelling to be significantly predictive of shunt malfunction, whereas other symptoms were not reliable. In our study we found a palpable abdominal cyst to be very predictive and specific for abdominal dislocation. This sign was not found in any other diagnosis. All other symptoms were not specific enough to distinguish between each diagnosis and therefore to guide further invasive radiological investigations.

Hence we think our diagnostic devices or at least pathways have to be improved to prevent these patients from undergoing needless examinations.

The most important finding in our study was a time dependency of certain shunt complications. Using recursive partition analysis, we could divide shunt complications according their probability of occurrence into three time periods. Early complications occurring before 6 months were abdominal dislocations and infections. Complications predominantly appearing within 6 months to 4 years were over- and underdrainage. Late complications such as kinking, laceration and disconnections presented particularly after 4

years. However, blocking of the shunt was with 20% to 25% of cases a constantly occurring complication after 6 months.

Based on these frequencies we developed a decision tree leading to a more focused and therefore less invasive diagnostic workup. This statement has of course to be proven by a prospective study, which was started in our department.

We are aware of the limitations of the presented study. First of all, there is the retrospective character, which leads to many unanswered questions. The limited number of patients lead to a low number of rare shunt dysfunctions such as abdominal dislocations, over-drainage or shunt infections which made further analysis difficult. Some rare complications might be not detected in a limited number of cases. We therefore think that a replication of our results and further testing of our suggested diagnostic workup in other cohorts of patients would be reasonable.

The data, which could be derived from our patient files, was not sufficient to provide us with a diagnosis in all cases in which no SD was found. Furthermore, symptoms were not recorded in a standardized manner and therefore conclusions regarding symptom constellations being specific for certain diagnoses were impossible.

There might even be some differences in SD manifestation depending on the underlying illness which lead to initial shunt implantation. We did not consider this in our study since patient population would be even more divided in to subgroups and statistics would become less reliable. Second, other previous studies already failed to find specific symptoms that would change the amount of further examinations and the latter was the main aim of our study.

Similarly the fact, that some shunt systems offer the possibility to gain some information about the shunt function by pumping the shunt chamber was not investigated in this study because of heterogeneity of the shunt systems. The Specificity and Sensitivity of this clinical test is unknown and might be worth to investigate in future more valve type focused studies.

Last, a very large portion of the SD was over- and underdrainage. Modern shunt systems mostly include valves which prevent the so-called siphoning-effect, which can lead to over-drainage and consequently to very careful adjustment of the differential-pressure-valves to relatively high pressure levels, meaning underdrainage. At our department we introduced these modern valve systems systematically in 2014 and we seem to see only few over- and underdrainage complications since. A prospective study will therefore provide a more actual view on the referred shunt-dysfunctions.

To the bets of our knowledge, time dependencies of complications have not been published till now. We are confident that the proposed decision tree will be helpful optimizing diagnostics in these situations and will motivate other workgroups to try further refinement.

Conclusions

Admission for suspected SD are very frequent. Only 46% of all admissions will have a real shunt related problem. Since symptoms and signs are very unspecific further invasive radiological examinations remain an indispensable part of the diagnostic workup. The awareness of a time dependency of specific shunt complications can help to focus the search for the cause of a SD and therefore lower the invasiveness of the diagnostic procedures.

Funding: No funding was received for this research.

Conflict of Interest: All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Ethical approval: All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: For this type of study formal consent is not required.

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Figures

Tables

Figure 1

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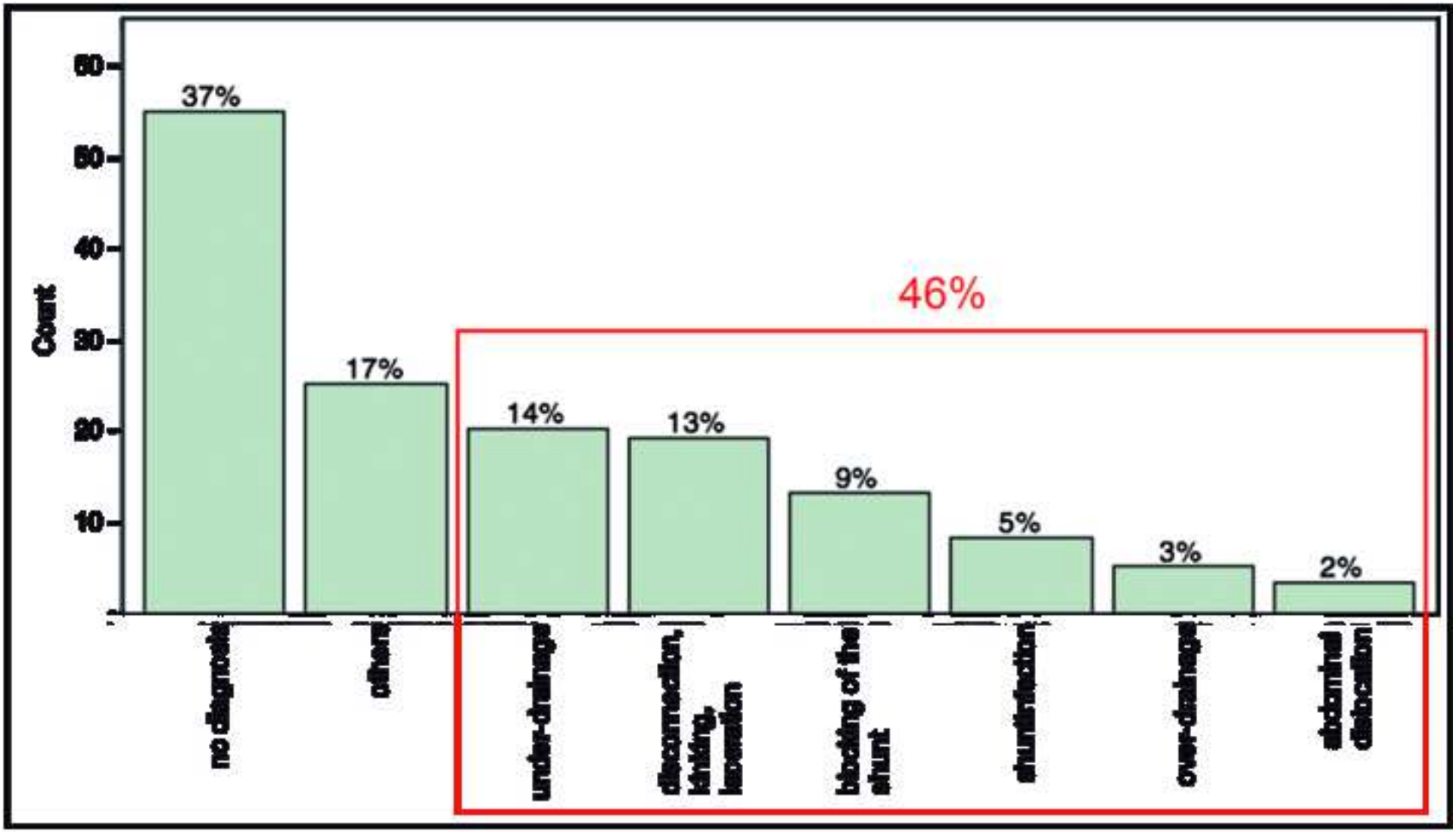

Figure 2

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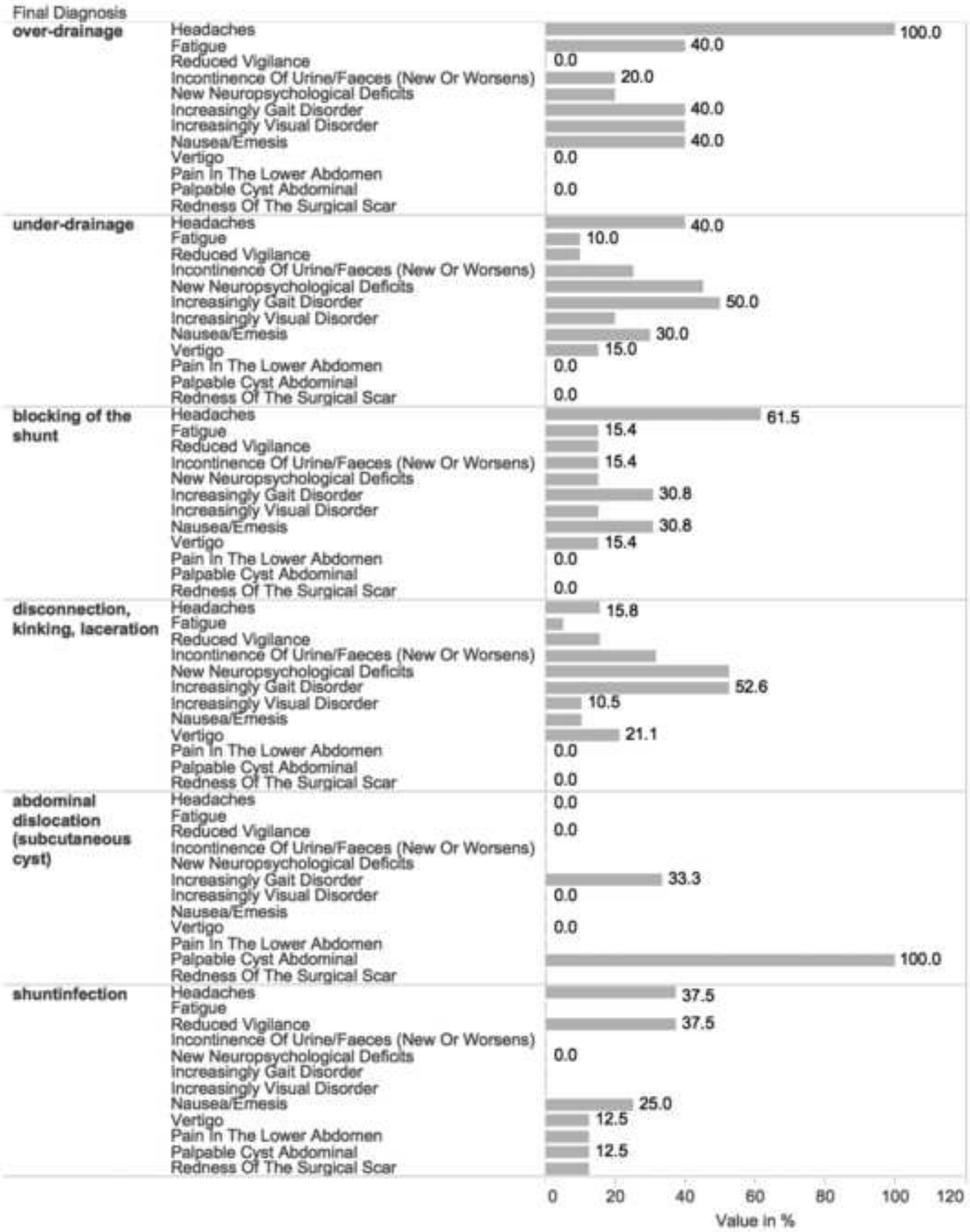

Figure 3

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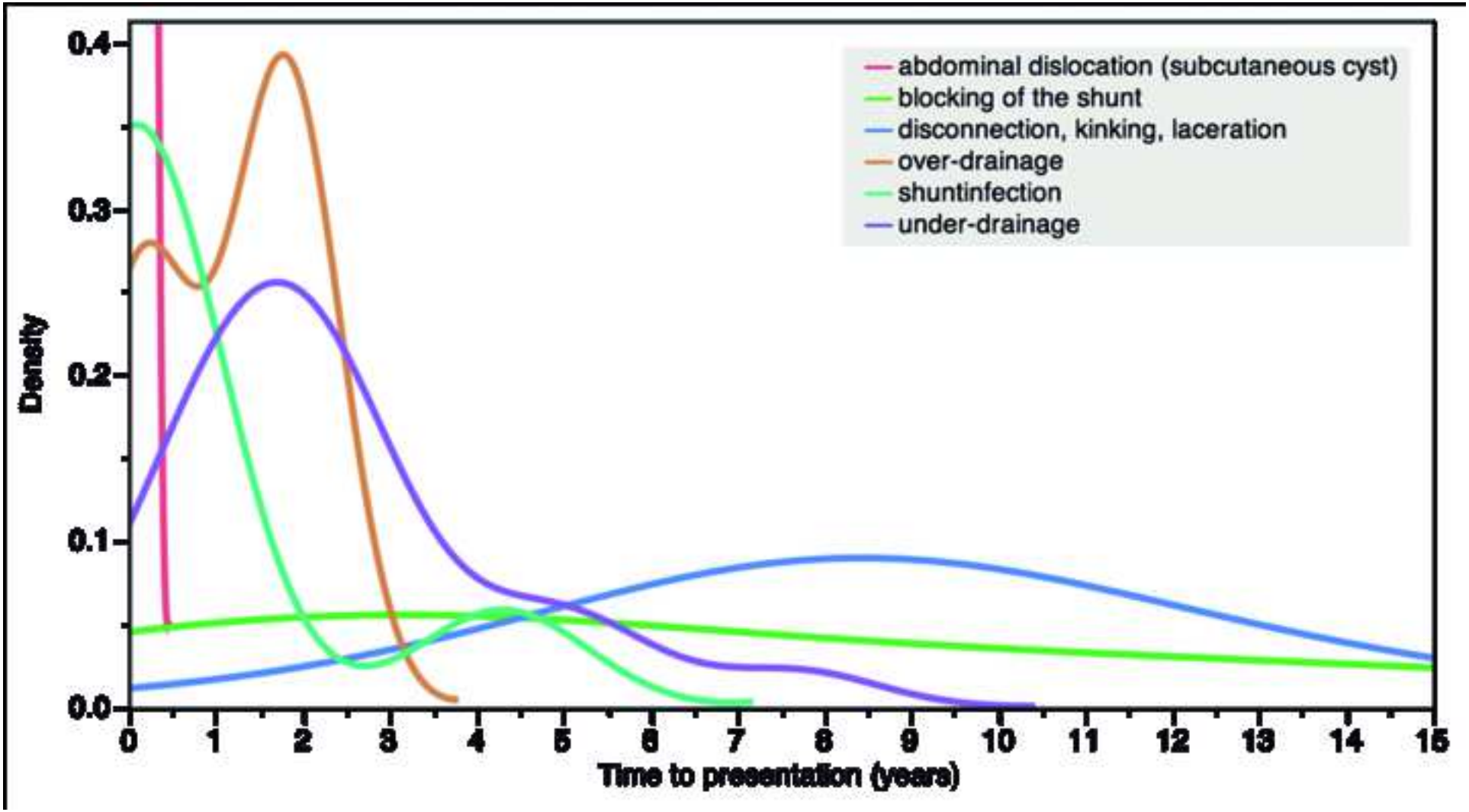

Figure 4

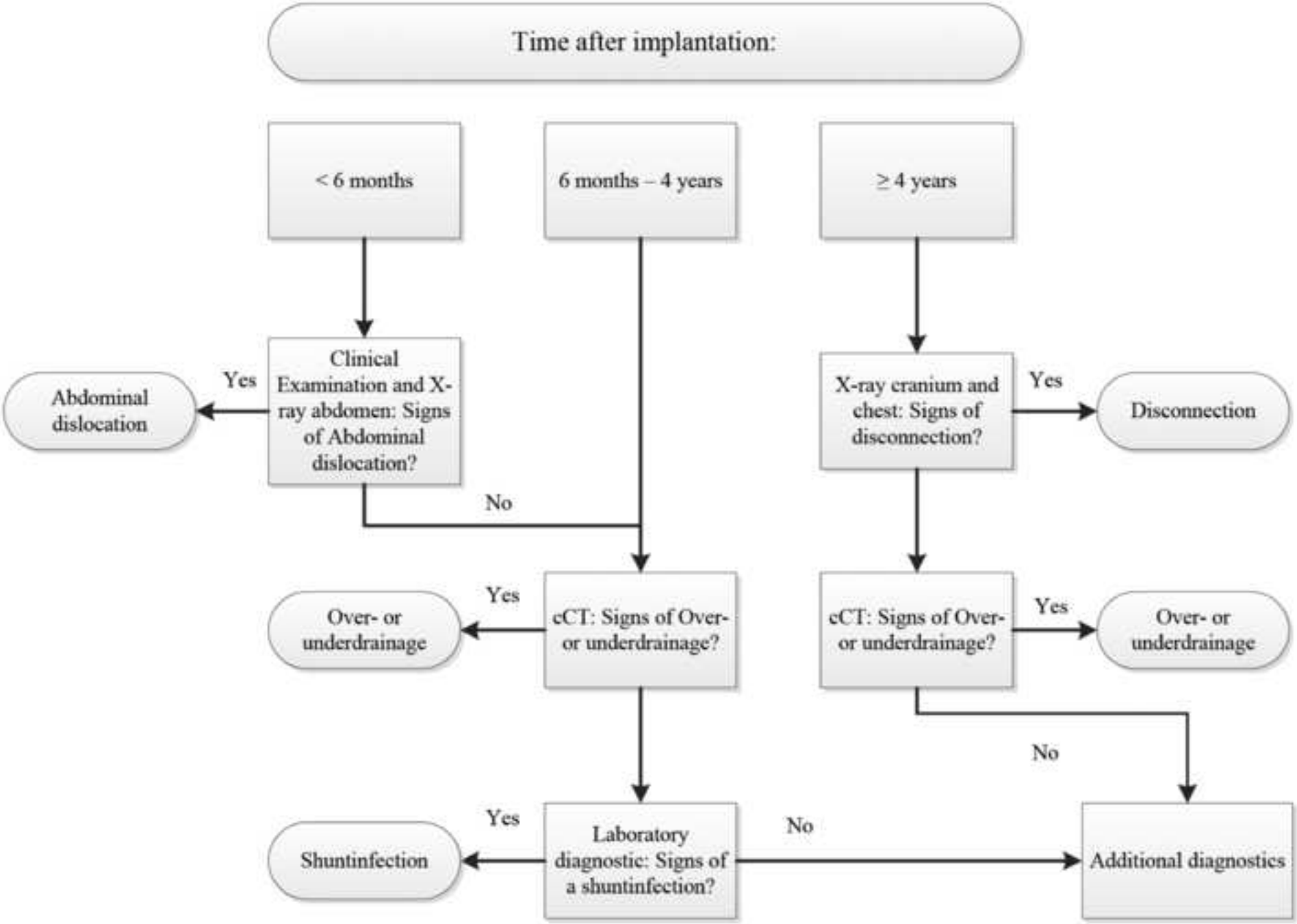

Table 1

Referrer	n	% of total
general practitioner	45	30.4%
rehab or other hospital	37	25.0%
patient self-admission	36	24.3%
neurologist	22	14.9%
nursing home	4	2.7%
not specified	2	1.4%
otorhinolaryngologists	1	0.7%
psychiatrist	1	0.7%

Table 2

Diagnostic procedure	n	% of total
CCT native	128	86%
CCT with contrast agent	86	58%
x-ray cranium	88	59%
x-ray thorax	91	61%
x-ray abdomen	81	55%
shunt tap	17	11%

Table 3

Type of Shunt dysfunction	Prob <6 months	Prob 6 months - 4 years	Prob >4 years
abdominal dislocation	22%	0%	0%
over-drainage	15%	13%	0%
shuntinfection	44%	0%	6%
under-drainage	16%	60%	14%
blocking of the shunt	1%	22%	25%
disconnection, kinking, laceration	2%	5%	55%

Table 4

Type of shunt dysfunction	Area Under the ROC Curve
abdominal dislocation	0.9095
over-drainage	0.7558
shuntinfection	0.8482
under-drainage	0.753
blocking of the shunt	0.6263
disconnection, kinking, laceration	0.8203